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MODERN CONCEPTS OF CARDIOVASCULAR DISEASE

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Clinical Features and Diagnosis of Primary Myocardial Disease (I) *†

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Clinical interest in diseases which primarily involve the myocardium has waxed and waned over the years. Before the period of clinical recognition of coronary artery disease and its secondary myocardial complications, many poorly understood and unexplained illnesses and cardiac deaths were diagnosed as "chronic myocarditis or myocardial degeneration." These were the most commonly recorded causes of cardiac deaths in adults in the United States during the first two decades of the present century. Following the recognition of the clinical features of coronary disease, noncoronary myocardial disease became essentially an occasional pathological diagnosis, except for that clinically recognized as a part of the cardiac lesions in rheumatic fever in children and young adults and myocarditis associated with diptheria and a few viral and rickettsial diseases.

Christian,[1]‡ in 1950, stressed the importance of the myocardium, its great vulnerability on the basis of its function and structure in relation to injurious agents, and the importance of understanding the mechanisms of dilatation, hypertrophy, and muscle failure that are so characteristic of the subacute and chronic forms of myocardial disease. He considered the recognition of myocardial insufficiency to be of clinical importance and defined myocardial insufficiency as "a condition in which there is evidence during life of failure of the heart muscle to function with normal efficiency." When one sums up the numerous faults that can occur in the function of the efficient myocardium, one will have the clinical features as presented by primary disease of the myocardium, namely: failure to function as an efficient pump in supplying the coronary, systemic and pulmonary circulations (congestive and anginal failure), failure to initiate and conduct a normal heart beat (arrhythmias, conduction disorders, sudden deaths), and failure to maintain muscle tone and support the valvular structures (dilatation, hypertrophy, incompetency of valves).

The diseases which may be responsible for these faults in myocardial function are numerous and diversified in nature. In a broad sense, they can be divided into two major categories: (1) primary myocardial diseases; and (2) circulatory diseases which have their primary lesions elsewhere in the cardiovascular system, with a secondary effect on the myocardium.

The term "primary myocardial disease (PMD)," as used in this publication and previous publications,[2, 3, 4] refers to those diseases of diverse etiology which specifically and primarily involve the myocardium rather than

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† Part II of this article will appear in the September 1961 issue.

‡ References for Parts I and II of this article will appear at the end of Part II.

TABLE 1

ETIOLOGICAL AND CLINICAL FORMS OF PRIMARY MYOCARDIAL DISEASE

Myocarditis, acute, subacute, and chronic:
Myocarditis due to infectious etiological agents
Idiopathic myocarditis
Myocarditis of collagen vascular diseases
Myocarditis of physical and metabolic origin
Obscure myocardial diseases (probably healed myocarditis):
Fibroelastosis
Endomyocardial fibrosis
Idiopathic cardiac hypertrophy
Familial cardiomegaly
Myocardial diseases associated with systemic diseases:
Amyloidosis
Hemochromatosis
Sarcoidosis
Carcinomatosis
Anemias
Glycogen storage disease
Friedreich's ataxia
Progressive muscular dystrophy
Primary myocardial tumors

other areas of the cardiovascular system (table 1). Other publications have referred to these diseases by such terms as cardiomyopathies,[5, 6, 7] uncommon forms of myocardial disease, noncoronary myocardial degeneration, or simply by such general terms as myocarditis or myocardosis,[8] depending upon whether or not an element of infection is present. Others, in their clinical and pathological evaluation, have simply chosen to refer to such myocardial diseases as being noncoronary, nonrheumatic, or nonhypertensive myocardial disease. More chronic, insidious, and poorly understood forms of PMD, some with familial characteristics, have been reported under the classification of familial cardiomegaly,[9] idiopathic myocardial hypertrophy,[10] and asymmetrical hypertrophy.[11] Where the chronic myocardial disease has been associated with secondary changes in the endocardium, they have been reported under various entities such as fibroelastosis,[12] myocarditis with endocardial elastomyofibrosis,[13] or endomyofibrosis.

The clinical features of myocardial insufficiency secondary to the more common forms of heart disease, such as obliterative coronary atherosclerosis, systemic and pulmonary hypertensive vascular disease, long-standing systolic or diastolic overload of the heart resulting from congenital and acquired valvular lesions, intra- and extracardiac shunts, obstructive vascular lesions or constrictive pericardial disease, are well recognized and diagnosed today. Recognition of these forms of secondary myocardial disease is greatly aided by our understanding of the clinical features of the primary lesions elsewhere in the circulatory system.

Early clinical diagnosis, or even an ante mortem diagnosis of the group of diseases included as primary myocardial diseases, have been infrequently made. Great difficulty has existed in the establishment of an acceptable clinical picture and criteria for clinical diagnosis. Too often, the clinical diagnosis, when made, is established by exclusion of other common causes of myocardial insufficiency or by the presence of nonspecific T wave changes in the electrocardiogram. The great majority of case reports have come from pathologists or from clinicians' retrospective review of clinical features. The purpose of this paper is to present the clinical features which, from clinical experience, have permitted an early diagnosis and a positive diagnosis of PMD, rather than a diagnosis by exclusion of other forms of heart disease.

In spite of the great diversity and multiplicity of causes and the variations in the clinical course, as presented by the primary forms of myocardial disease, it has been observed that they present clinical features which permit a clinical diagnosis with an accuracy approaching that of the more common forms of cardiovascular disease.[2] The most commonly observed clinical features have been as follows: congestive failure, cardiomegaly, arrhythmias and conduction disorders, auscultatory abnormalities, or combinations of the above.

GENERAL FEATURES

PMD is seen in all age groups. Coxsackie viral myocarditis occurs in the newborn. Acute, subacute and chronic myocarditis of specific and unknown etiology is seen throughout life as a frequent complication of systemic diseases.[14] Subacute myocarditis, as well as probable chronic and healed forms, reported as idiopathic cardiomegaly and various forms of endomyocardial fibrosis, have a high incidence in young adults but again are seen at all ages. Multiple cases have occurred in families and have been reported as familial cardiomegaly. Forms of PMD associated with systemic diseases likewise occur in all age groups. For example, myocardial involvement with myocardial insufficiency is seen

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MORNING	SESSIONS ON CLINICAL CARDIOLOGY	Address by President Conner Memorial Lecture Symposium: Contribution of Phonocardiography to Auscultation	Panel: Ventricular Arrhythmias Closed Chest Cardiac Resuscitation	Jointly with Basic Science Symposium: Role of Hormones in Heart Failure Venous Factors in Control of the Circulation
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	SPECIAL AND COUNCIL SESSIONS	ARTERIOSCLEROSIS	BASIC SCIENCE	RHEUMATIC FEVER and CONGENITAL HEART DISEASE
		CIRCULATION	CARDIOVASCULAR SURGERY Jointly with RHEUMATIC FEVER and CONGENITAL HEART DISEASE	COMMUNITY SERVICE and EDUCATION Session for Public
		COMMUNITY SERVICE and EDUCATION Session for Nurses	COMMUNITY SERVICE and EDUCATION COMPENSABLE HEART DISEASE, STRAIN AND TRAUMA	CARDIOVASCULAR FILMS
	EVENING		Cardiac Conferences	Annual Dinner American Heart Association

Sunday, October 22 — 2:00 P.M.	Program for Assembly Delegates
Monday, October 23 — all day	Assembly Panel Meetings
12:15 P.M.	Assembly Luncheon
Tuesday, October 24 — 9:00 A.M. — 12 Noon	Annual Meeting of Assembly, elections — panel reports.

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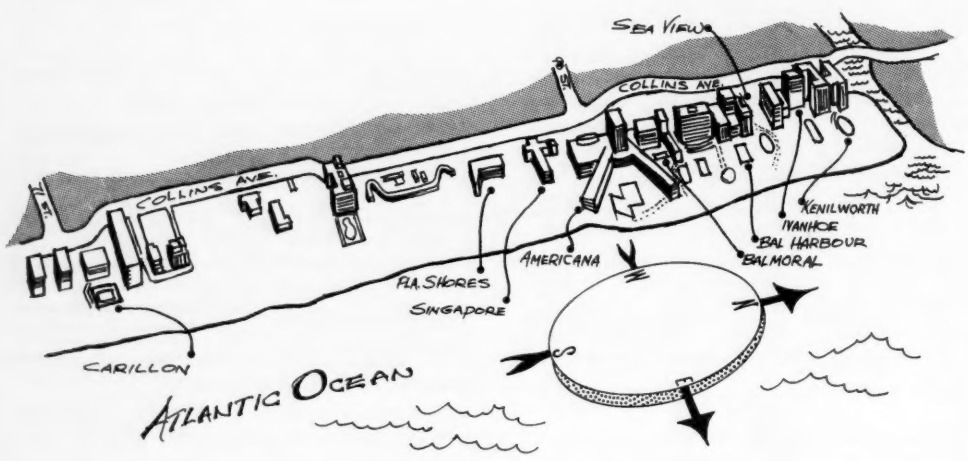

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The following special events have been planned. More detailed information will be available later.

FRIDAY, OCTOBER 20

10:00 A.M.	Opening of the Heart Hospitality Room. Special Open House from 3:00 to 5:00 P.M. for meeting and greeting friends.

SATURDAY, OCTOBER 21

12:30 P.M.-3:00 P.M.	Luncheon and Fashion Show at the fabulous Fontainebleau Hotel, Miami Beach.

SUNDAY, OCTOBER 22

12:30 P.M.-3:30 P.M.	Scenic boat trip around beautiful waterfront residences—including buffet luncheon on board boat.

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in the infant in glycogen storage disease. Primary myocardial involvement occurs in gargoylism in children. Myocardial fibrosis, hypertrophy and failure occur in the various forms of neuromuscular diseases, such as Friedreich's ataxia and progressive muscular dystrophy, which are seen primarily in childhood and young adults. Amyloidosis of the primary type, or that seen in myeloma, frequently involves the myocardium and constitutes a form of PMD which is rarely seen under the age of 40 years. Review of the literature and personal experience indicates that at least a part of the so-called postpartum heart disease consists of forms of PMD.

The incidence of myocarditis, the most important form of PMD as based on clinical recognition, is definitely lower than the pathological incidence; the latter is grossly inaccurate, as the myocardium is generally inadequately studied histologically and totally neglected from a histochemical, biochemical, and hemodynamic standpoint in routine autopsies. The autopsy incidence of myocarditis is given as about 3.5 per cent of all cases in routine studies, and Saphir[15] states that it rises to about 8 to 10 per cent if many sections are cut from the myocardium. This is much higher than the clinical diagnosis. The discrepancy between the clinical and pathological incidence has been ascribed to two factors:[16] the relatively innocent nature of the clinical and laboratory findings and, in many cases, the apparent reluctance of the clinician to make the diagnosis.

SYMPTOMATOLOGY AND CLINICAL COURSE

The symptomatology and clinical course of PMD is variable. When myocarditis occurs as a part of a known infectious disease, or fibrosis or other degenerative changes occur in the myocardium during the course of a systemic disease other than that of the cardiovascular system, the cardiac symptomatology is frequently overshadowed or masked by that of the primary disease, or in case of the infectious disease, the symptoms develop insidiously after recovery from the primary disease. Idiopathic, or so-called isolated forms of myocarditis, likewise may have an abrupt onset, simulating an acute myocardial infarction, or the onset may be so insidious that sudden death may occur without recognition of illness. Subacute forms of both specific and nonspecific myocarditis exist which present a slow but relentless and progressive downhill course lasting months to several years and have been referred to as pernicious myocarditis.[17] Chronic forms of myocardial disease, with symptoms over years and reported as idiopathic cardiomegaly,[4, 10] etc., which at autopsy show little evidence of residual myocarditis or fibrosis, in my opinion represent instances where the myocarditis heals without fibrosis and sclerosis but the hemodynamic and mechanical faults of irreversible dilatation and compensating hypertrophy finally result in death from muscle failure. Others in this chronic group will develop secondary changes in the endocardium of varying degrees with fibrosis and elastosis of both the myocardium and endocardium. This latter process is a complication of other forms of primary and secondary myocardial diseases and is in itself not a separate disease.[18, 19] Complicating thromboembolism to the pulmonary and systemic circulations and intractable cardiac failure usually terminate the course of the chronic cases, the duration of which is variable, often lasting years.

The symptoms of PMD include many that are common to other forms of heart disease, and when considered alone they are neither distinctive nor diagnostic. However, when considered in conjunction with their mode of onset, their clinical course, environmental factors, and associated physical signs, they are helpful in diagnosis.

Weakness, fatigue, and exertional breathlessness, are common early manifestations of myocardial insufficiency, often occurring unnoticed by the physician long before frank congestive features develop in the pulmonary and systemic circulations. In the absence of overt signs of heart disease, iatrogenic heart disease and psychosomatic diagnoses are commonly made as an early diagnosis. Later, when obvious features of congestive failure develop, weakness and fatigue continue as predominant symptoms, out of proportion to that seen in other forms of heart disease. This is the result of the low and often relatively fixed cardiac output seen in advanced PMD. Symptoms of biventricular failure often occur simultaneously or follow in rapid succession. This feature is more distinctive of the failure of PMD than of failure seen in other forms of heart disease.

Dizziness, lightheadedness, and episodes of exertional syncope, or syncope at rest, occur frequently and may be an early and predominant symptom. Again, the absence of overt cardiac abnormalities often result in a noncardiac or functional diagnosis. Taken alone, they cannot be distinguished from such symptoms occurring from low cardiac output in pa-

tients with obstructive valvular lesions, pulmonary or systemic hypertensive vascular disease, or from ectopic rhythms and conduction disorders associated with other forms of heart disease. Correlation with other findings is important. Sudden death, from serious ectopic rhythms or conduction disorders, is common in both the acute and chronic forms, but especially in acute myocarditis.

Precordial pain is not common, and when present is usually the result of relative coronary insufficiency due to a low cardiac output, to pulmonary hypertension associated with the failure, and occasionally to an associated pericarditis. Pain or discomfort in the upper right quadrant associated with congestive hepatomegaly is far more common and can be very distressing, especially that which may occur with episodes of acute right ventricular dilatation and tricuspid incompetency. Chest pain and pain in splenic and renal areas occur when pulmonary and systemic embolization are frequent complications in chronic forms of the disease. Symptoms of combined pulmonary and systemic embolization warrant consideration of the presence of thrombi in chambers of both sides of the heart. Primary myocardial disease is a more common cause of biventricular mural thrombi than either coronary disease or chronic valvular heart disease.

The general physical condition of the patient at the time of evaluation may vary from that of apparent excellent health, to the discovery of cardiomegaly and bundle-branch block on routine annual physical examination (as occurred in the case of a young resident of the house staff who died of chronic myocarditis less than three years later), to that of an acutely or chronically ill, bedridden patient with intractable congestive failure. The specific physical findings will be described under subsequent headings, but some general features may be enumerated.

Cyanosis, if present, is that of stasis cyanosis related to a poor circulation. With a markedly reduced cardiac output, and in the presence of venous congestion, the resulting cyanosis may be so pronounced as to lead to an erroneous impression of congenital lesions with right to left shunts.

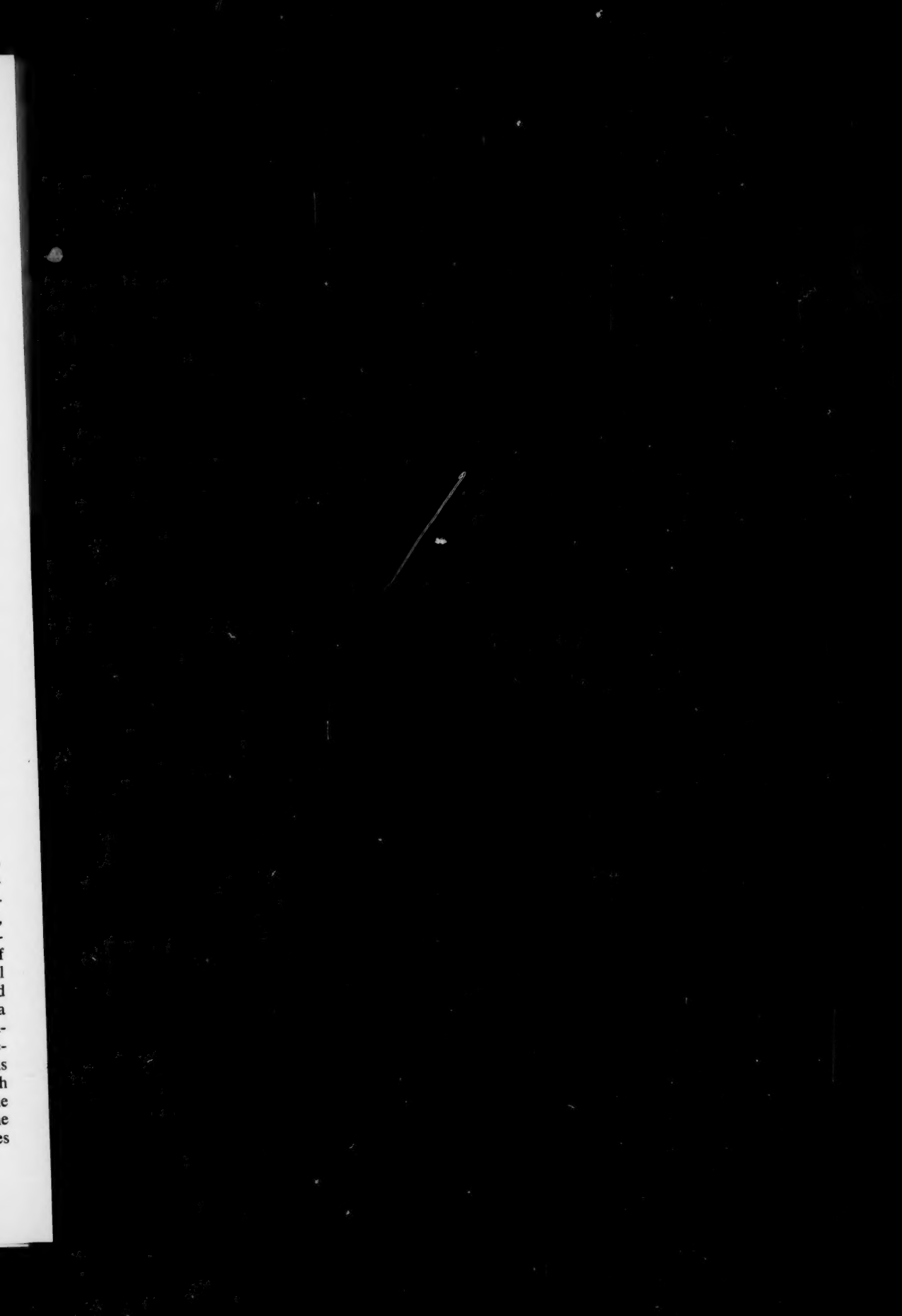
Persistent elevation of arterial pressure is not a part of the finding in PMD. Transitory hypertension may exist for variable periods of acute failure. If persistent, one should look for some complication, such as associated renal disease from complicating thrombo-embolism, scleroderma, lupus, or other causes. The arterial pulse is usually small, and with progressive failure the systolic pressure is often low and the pulse pressure narrow.

Varying degrees of peripheral edema may be present. Pleural effusion occurs and is often related to pulmonary infarctions. Hepatomegaly is often pronounced and persistent and pulsations of the liver are frequent because of the common occurrence of elevated venous pressure and tricuspid valve incompetence. The neck veins are frequently prominent as a result of persistent elevation of venous pressure, and in those instances where tricuspid incompetency is present, prominent pulsations are easily seen.

CARDIOMEGALY AND OTHER ROENTGENOLOGICAL FINDINGS

The size of the heart, as determined by clinical and roentgenological examinations, may fall within normal limits in a few patients seen early, or where restrictive fibrosis prevents dilatation, or if seen following recovery from cardiac dilatation. However, in the majority, gross cardiomegaly is readily recognizable by clinical examination and roentgenological studies. Except in acute forms of PMD, such as acute fatal myocarditis, this cardiomegaly is the result of combined chamber dilatation and interstitial fibrosis, endocardial sclerosis, and muscle hypertrophy. Isolated cardiac dilatation is recognized clinically by a quiet precordium and an enlarged, globular cardiac silhouette on physical examination, chest roentgenogram, and fluoroscopy, but unassociated with electrocardiographic features of either isolated right or left, or combined ventricular hypertrophy. These features frequently suggest pericardial disease with effusion.

The cardiomegaly of chronic form is usually that of a generalized chamber enlargement with elements of both dilatation and hypertrophy. With the development of failure, progressive dilatation of the ventricular chambers results in dilatation and incompetency of the atrioventricular valves. Following mitral and tricuspid valve incompetencies, right and left atrial enlargement develops, often to a pronounced degree. These radiological features, plus the murmurs of valvular incompetence, often result in an erroneous diagnosis of organic valve disease. An observation which has had value in differential diagnosis is the pronounced change in cardiac size with the development of, and recovery from, episodes



of failure, along with a decrease or disappearance of the murmurs as the heart size and failure improves. This is usually the opposite in dealing with organic valve disease, where the murmurs are heard with maximum intensity when the heart is small and compensated and absent or decreased in intensity when the heart is dilated and in failure. In a chronic case with slowly developing hypertrophy, the enlargement is frequently that of isolated left ventricular hypertrophy, and in those with asymmetrical forms of hypertrophy of the ventricular chambers, the outflow tract of the left ventricle is most often involved, thus simulating organic aortic or pulmonic stenosis. Again, as failure develops, as in the asymmetrical type, generalized enlargement from dilatation occurs as in other forms. I have not seen calcification in the myocardium in roentgenological examinations, and it has not been reported by others. Special technics, such as angiocardiography and radioisotope scanning,[20] have been helpful in differentiating pericardial disease with effusion from an enlarged cardiac silhouette associated with PMD. Central streaming of contrast media in a dilated heart chamber may lead to an erroneous interpretation of increased width of the pericardial shadow in the angiocardiogram and lead to an erroneous diagnosis of pericardial effusion or thickening. In those forms of PMD with restrictive myocardial fibrosis and a normal size heart, the fluoroscopic and other roentgenographic findings offer little in differential diagnosis from constrictive pericardial disease except that calcification, when present, usually indicates calcified pericardium.

Other radiological findings, such as pleural effusions, pulmonary infarctions, pulmonary patterns of congestive failure, and pulmonary hypertension, occur at times but are no different from that seen with chronic congestive failure in other cardiovascular diseases. The combination of gross cardiomegaly, clear lung fields, and pronounced hepatomegaly, is a common finding in those with long-standing failure in whom tricuspid incompetency is present.

ARRHYTHMIAS, CONDUCTION DISORDERS AND ELECTROCARDIOGRAPHIC ABNORMALITIES

Arrhythmias and conduction disorders are usually observed at some time during the clinical course, especially as a terminal event, and are often responsible for a sudden and unexpected death. Ectopic rhythms of supraventricular, ventricular and nodal origin are seen, and the frequency, duration and response to therapy vary greatly. Ectopic premature contractions occurring at intervals between bouts of tachycardia, may provide a clue as to the site of origin of the arrhythmia not recorded by an electrocardiogram. A sinus tachycardia out of proportion to any existing fever, not associated with an endocrine disorder or other systemic disease, and with clinical features of mild failure, should cause one to consider myocardial disease as the cause of the tachycardia. This has been a clinical feature of importance in recognizing myocarditis associated with infectious diseases.[16]

Intraventricular and atrioventricular conduction defects of some degree are usually manifested sometime during the clinical course. Complete A-V dissociation occurs at times: it may be a terminal event, or it may exist for years before death. The intraventricular conduction disturbances are more common, and are often characterized by an intraventricular conduction delay or block of an indeterminate type, or by shifting patterns of block of the right and left bundle systems. The latter abnormality should cause a strong suspicion of PMD, even in older adults where coronary disease is the cause of more frequent secondary types of myocardial disease. Likewise, the presence of an otherwise unexplained bundle-branch block, especially if it is of the left bundle type in a young individual, even if asymptomatic or intermittent, should alert the physician to look for other features of PMD. Functional bundle-branch block does occur in the absence of myocardial disease, but is rare. The frequency of a persistent bundle-branch block as a complication of chagasic and diphtheritic myocarditis are classical examples of the relationship of conduction disorders to PMD. I have likewise seen two instances of classical accelerated A-V conduction occurring in fatal cases of myocarditis. This has been observed by others.[21]

There are other electrocardiographic features that are common in, but not diagnostic of, PMD. The most important and reliable are combinations of abnormalities which indicate diffuse myocardial involvement, such as combination of low voltage, A-V or I-V conduction disturbances, or arrhythmias. Low voltage alone may occur in such confusing conditions as pericarditis with effusion, but the association of arrhythmias and conduction disorders are uncommon in pericarditis unless there is an associated myocardial disease. Much has been

published on the S-T segment and T wave abnormalities in myocardial disease but these abnormalities alone are not diagnostic of PMD and have resulted in erroneous diagnosis. These nonspecific ST-T changes are of greatest diagnostic value when observed to develop in serial electrocardiograms during the course of an illness, such as mumps, diphtheria, etc. In such instances, these changes may be used as a reliable index of some intramural damage by an existing myocarditis, but such changes are not always reliable as evidence of chronic myocardial disorders. Similar serial changes are helpful in following instances of toxic myocarditis associated with drug therapy, such as that occurring with digitalis, emetine, etc., or myocardial damage by other chemicals or physical agents, including trauma. There is a tendency, however, to place too much reliance on this type of ECG change to the exclusion of the total clinical evaluation. The electrocardiogram is likewise helpful in confirming the state of hypertrophy and dilatation.

AUSCULTATORY FINDINGS

The most frequent abnormalities found on auscultation are the result of myocardial insufficiency with the production of gallop rhythm and dilatation of the heart chambers and the atrioventricular valve rings with resultant murmurs of mitral and tricuspid incompetency. The second heart sound, as heard in the pulmonary valve area, is frequently accentuated as the result of pulmonary hypertension and occasionally there may be a diastolic murmur of pulmonary valve incompetency in this same area from the same cause. Ectopic rhythms and conduction disorders, when present, are productive of additional auscultatory abnormalities of the heart sounds. When hypertrophic muscular changes occur in the outflow tracts of the ventricles, obstructive systolic murmurs develop which cannot be differentiated by auscultation alone from obstructive organic valve lesions. An occasional complicating pericarditis adds additional auscultatory findings. The origin and nature of the varied auscultatory abnormalities have been well described and illustrated by Harvey.[3, 22]

The most consistent auscultatory finding in PMD, regardless of the type and etiology, is the presence of a diastolic gallop rhythm. It is present in essentially all instances where failure is present, and at times its presence may precede overt symptoms and other signs of failure. Ventricular gallop is more common, but atrial gallops occur as well. At times both may be present, especially in chronic forms of the disease. Burchell,[23] in his discussion of the diagnosis of unusual causes of heart failure, states that the diagnosis of PMD should be considered when a patient with heart disease has ventricular and/or atrial gallop, cardiomegaly, and other signs of cardiac decompensation.

A systolic murmur of variable quality, often reaching grade IV, or even grade VI, intensity and pansystolic in duration, develops at the apical region during the observation of heart failure or may be present when the patient is initially examined. At times, the murmur is widely transmitted to the base of the heart, to the axilla and left back, and simulates an organic mitral or aortic valve lesion, or is confused with murmurs of congenital malformations. This murmur is usually the result of mitral valve incompetency. When right ventricular failure and dilatation are pronounced, a systolic murmur with inspiratory accentuation characteristic of tricuspid valve incompetency may develop. Usually the presence of systolic venous pulsations in the neck veins and/or a pulsating liver makes the diagnosis of tricuspid incompetency more evident. The unstable and changing character of these murmurs, with frequent disappearance following recovery or improvement from an episode of failure and cardiac dilatation only to reappear in another bout of acute failure or progression of the failure with increased activity, are important auscultatory features which are valuable in the differential diagnosis of PMD with valvular incompetencies and primary valvular heart disease with failure. In the latter, the murmurs often decrease during a bout of failure and are most pronounced in the interval between bouts of failure.

EMBOLIC FEATURES

Thrombo-embolism, with thrombi originating in the peripheral veins, and with emboli going to the lungs, is common to many forms of heart disease and circulatory disorders. This may also occur in PMD. The development of mural thrombi in the ventricular and atrial chambers is frequent in the subacute and chronic forms of the disease. Embolization to both the pulmonary and systemic circulations occur from this source. Thus the presence of multiple episodes of embolism to pulmonary and systemic areas in a patient with cardiomegaly and failure of unknown cause should arouse a suspicion of PMD.